



## SEQUENCE LISTING

<110> Riordan, John R.  
Chang, Xiu-bao

<120> CFTR POLYPEPTIDES, FRAGMENTS THEREOF AND  
METHODS OF USE TO OVERCOME BIOSYNTHETIC MISPROCESSING

<130> 07039-167002

<140> US 10/030,585

<141> 2002-05-22

<150> PCT/US00/40324

<151> 2000-07-07

<150> US 60/143,090

<151> 1999-07-09

<150> US 60/146,097

<151> 1999-07-21

<160> 17

<170> FastSEQ for Windows Version 4.0

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Glu	Leu	Ser	Asp	Ile	Tyr	Gln	Ile	Pro	Ser	Val	Asp	Ser	Ala	Asp	Asn	35	40	45	
Leu	Ser	Glu	Lys	Leu	Glu	Arg	Glu	Trp	Asp	Arg	Glu	Leu	Ala	Ser	Lys	50	55	60	
Lys	Asn	Pro	Lys	Leu	Ile	Asn	Ala	Leu	Arg	Arg	Cys	Phe	Phe	Trp	Arg	65	70	75	80
Phe	Met	Phe	Tyr	Gly	Ile	Phe	Leu	Tyr	Leu	Gly	Glu	Val	Thr	Lys	Ala	85	90	95	
Val	Gln	Pro	Leu	Leu	Leu	Gly	Arg	Ile	Ile	Ala	Ser	Tyr	Asp	Pro	Asp	100	105	110	
Asn	Lys	Glu	Glu	Arg	Ser	Ile	Ala	Ile	Tyr	Leu	Gly	Ile	Gly	Leu	Cys	115	120	125	
Leu	Leu	Phe	Ile	Val	Arg	Thr	Leu	Leu	Leu	His	Pro	Ala	Ile	Phe	Gly	130	135	140	
Leu	His	His	Ile	Gly	Met	Gln	Met	Arg	Ile	Ala	Met	Phe	Ser	Leu	Ile	145	150	155	160
Tyr	Lys	Lys	Thr	Leu	Lys	Leu	Ser	Ser	Arg	Val	Leu	Asp	Lys	Ile	Ser	165	170	175	
Ile	Gly	Gln	Leu	Val	Ser	Leu	Leu	Ser	Asn	Asn	Leu	Asn	Lys	Phe	Asp	180	185	190	
Glu	Gly	Leu	Ala	Leu	Ala	His	Phe	Val	Trp	Ile	Ala	Pro	Leu	Gln	Val	195	200	205	
Ala	Leu	Leu	Met	Gly	Leu	Ile	Trp	Glu	Leu	Leu	Gln	Ala	Ser	Ala	Phe	210	215	220	
Cys	Gly	Leu	Gly	Phe	Leu	Ile	Val	Leu	Ala	Leu	Phe	Gln	Ala	Gly	Leu	225	230	235	240
Gly	Arg	Met	Met	Met	Lys	Tyr	Arg	Asp	Gln	Arg	Ala	Gly	Lys	Ile	Ser	245	250	255	
Glu	Arg	Leu	Val	Ile	Thr	Ser	Glu	Met	Ile	Glu	Asn	Ile	Gln	Ser	Val	260	265	270	
Lys	Ala	Tyr	Cys	Trp	Glu	Glu	Ala	Met	Glu	Lys	Met	Ile	Glu	Asn	Leu	275	280	285	
Arg	Gln	Thr	Glu	Leu	Lys	Leu	Thr	Arg	Lys	Ala	Ala	Tyr	Val	Arg	Tyr	290	295	300	
Phe	Asn	Ser	Ser	Ala	Phe	Phe	Phe	Ser	Gly	Phe	Phe	Val	Val	Phe	Leu	305	310	315	320
Ser	Val	Leu	Pro	Tyr	Ala	Leu	Ile	Lys	Gly	Ile	Ile	Leu	Arg	Lys	Ile	325	330	335	
Phe	Thr	Thr	Ile	Ser	Phe	Cys	Ile	Val	Leu	Arg	Met	Ala	Val	Thr	Arg	340	345	350	
Gln	Phe	Pro	Trp	Ala	Val	Gln	Thr	Trp	Tyr	Asp	Ser	Leu	Gly	Ala	Ile				

355	360	365
Asn Lys Ile Gln Asp Phe Leu Gln Lys Gln Glu Tyr Lys Thr Leu Glu		
370	375	380
Tyr Asn Leu Thr Thr Thr Glu Val Val Met Glu Asn Val Thr Ala Phe		
385	390	395
Trp Glu Glu Gly Phe Gly Glu Leu Phe Glu Lys Ala Lys Gln Asn Asn		400
	405	410
Asn Asn Arg Lys Thr Ser Asn Gly Asp Asp Ser Leu Phe Phe Ser Asn		415
	420	425
Phe Ser Leu Leu Gly Thr Pro Val Leu Lys Asp Ile Asn Phe Lys Ile		430
	435	440
Glu Arg Gly Gln Leu Leu Ala Val Ala Gly Ser Thr Gly Ala Gly Lys		445
	450	455
Thr Ser Leu Leu Met Met Ile Met Gly Glu Leu Glu Pro Ser Glu Gly		460
465	470	475
Lys Ile Lys His Ser Gly Arg Ile Ser Phe Cys Ser Gln Phe Ser Trp		480
	485	490
Ile Met Pro Gly Thr Ile Lys Glu Asn Ile Ile Phe Gly Val Ser Tyr		495
	500	505
Asp Glu Tyr Arg Tyr Arg Ser Val Ile Lys Ala Cys Gln Leu Glu Glu		510
	515	520
Asp Ile Ser Lys Phe Ala Glu Lys Asp Asn Ile Val Leu Gly Glu Gly		525
	530	535
Gly Ile Thr Leu Ser Gly Gly Gln Arg Ala Arg Ile Ser Leu Ala Arg		540
545	550	555
Ala Val Tyr Lys Asp Ala Asp Leu Tyr Leu Leu Asp Ser Pro Phe Gly		560
	565	570
Tyr Leu Asp Val Leu Thr Glu Lys Glu Ile Phe Glu Ser Cys Val Cys		575
	580	585
Lys Leu Met Ala Asn Lys Thr Arg Ile Leu Val Thr Ser Lys Met Glu		590
	595	600
His Leu Lys Lys Ala Asp Lys Ile Leu Ile Leu His Glu Gly Ser Ser		605
	610	615
Tyr Phe Tyr Gly Thr Phe Ser Glu Leu Gln Asn Leu Gln Pro Asp Phe		620
625	630	635
Ser Ser Lys Leu Met Gly Cys Asp Ser Phe Asp Gln Phe Ser Ala Glu		640
	645	650
Arg Arg Asn Ser Ile Leu Thr Glu Thr Leu His Arg Phe Ser Leu Glu		655
	660	665
Gly Asp Ala Pro Val Ser Trp Thr Glu Thr Lys Lys Gln Ser Phe Lys		670
	675	680
Gln Thr Gly Glu Phe Gly Glu Lys Arg Lys Asn Ser Ile Leu Asn Pro		685
	690	695
Ile Asn Ser Ile Arg Lys Phe Ser Ile Val Gln Lys Thr Pro Leu Gln		700
705	710	715
Met Asn Gly Ile Glu Glu Asp Ser Asp Glu Pro Leu Glu Arg Arg Leu		720
	725	730
Ser Leu Val Pro Asp Ser Glu Gln Gly Glu Ala Ile Leu Pro Arg Ile		735
	740	745
Ser Val Ile Ser Thr Gly Pro Thr Leu Gln Ala Arg Arg Arg Gln Ser		750
	755	760
Val Leu Asn Leu Met Thr His Ser Val Asn Gln Gly Gln Asn Ile His		765
	770	775
Arg Lys Thr Thr Ala Ser Thr Arg Lys Val Ser Leu Ala Pro Gln Ala		780
785	790	795
Asn Leu Thr Glu Leu Asp Ile Tyr Ser Arg Arg Leu Ser Gln Glu Thr		800
	805	810
		815

Gly	Leu	Glu	Ile	Ser	Glu	Glu	Ile	Asn	Glu	Glu	Asp	Leu	Lys	Glu	Cys	820	825	830
Phe	Phe	Asp	Asp	Met	Glu	Ser	Ile	Pro	Ala	Val	Thr	Thr	Trp	Asn	Thr	835	840	845
Tyr	Leu	Arg	Tyr	Ile	Thr	Val	His	Lys	Ser	Leu	Ile	Phe	Val	Leu	Ile	850	855	860
Trp	Cys	Leu	Val	Ile	Phe	Leu	Ala	Glu	Val	Ala	Ala	Ser	Leu	Val	Val	865	870	875
Leu	Trp	Leu	Leu	Gly	Asn	Thr	Pro	Leu	Gln	Asp	Lys	Gly	Asn	Ser	Thr	885	890	895
His	Ser	Arg	Asn	Asn	Ser	Tyr	Ala	Val	Ile	Ile	Thr	Ser	Thr	Ser	Ser	900	905	910
Tyr	Tyr	Val	Phe	Tyr	Ile	Tyr	Val	Gly	Val	Ala	Asp	Thr	Leu	Leu	Ala	915	920	925
Met	Gly	Phe	Phe	Arg	Gly	Leu	Pro	Leu	Val	His	Thr	Leu	Ile	Thr	Val	930	935	940
Ser	Lys	Ile	Leu	His	His	Lys	Met	Leu	His	Ser	Val	Leu	Gln	Ala	Pro	945	950	955
Met	Ser	Thr	Leu	Asn	Thr	Leu	Lys	Ala	Gly	Gly	Ile	Leu	Asn	Arg	Phe	965	970	975
Ser	Lys	Asp	Ile	Ala	Ile	Leu	Asp	Asp	Leu	Leu	Pro	Leu	Thr	Ile	Phe	980	985	990
Asp	Phe	Ile	Gln	Leu	Leu	Leu	Ile	Val	Ile	Gly	Ala	Ile	Ala	Val	Val	995	1000	1005
Ala	Val	Leu	Gln	Pro	Tyr	Ile	Phe	Val	Ala	Thr	Val	Pro	Val	Ile	Val	1010	1015	1020
Ala	Phe	Ile	Met	Leu	Arg	Ala	Tyr	Phe	Leu	Gln	Thr	Ser	Gln	Gln	Leu	1025	1030	1035
Lys	Gln	Leu	Glu	Ser	Glu	Gly	Arg	Ser	Pro	Ile	Phe	Thr	His	Leu	Val	1045	1050	1055
Thr	Ser	Leu	Lys	Gly	Leu	Trp	Thr	Leu	Arg	Ala	Phe	Gly	Arg	Gln	Pro	1060	1065	1070
Tyr	Phe	Glu	Thr	Leu	Phe	His	Lys	Ala	Leu	Asn	Leu	His	Thr	Ala	Asn	1075	1080	1085
Trp	Phe	Leu	Tyr	Leu	Ser	Thr	Leu	Arg	Trp	Phe	Gln	Met	Arg	Ile	Glu	1090	1095	1100
Met	Ile	Phe	Val	Ile	Phe	Phe	Ile	Ala	Val	Thr	Phe	Ile	Ser	Ile	Leu	1105	1110	1115
Thr	Thr	Gly	Glu	Gly	Glu	Gly	Arg	Val	Gly	Ile	Ile	Leu	Thr	Leu	Ala	1125	1130	1135
Met	Asn	Ile	Met	Ser	Thr	Leu	Gln	Trp	Ala	Val	Asn	Ser	Ser	Ile	Asp	1140	1145	1150
Val	Asp	Ser	Leu	Met	Arg	Ser	Val	Ser	Arg	Val	Phe	Lys	Phe	Ile	Asp	1155	1160	1165
Met	Pro	Thr	Glu	Gly	Lys	Pro	Thr	Lys	Ser	Thr	Lys	Pro	Tyr	Lys	Asn	1170	1175	1180
Gly	Gln	Leu	Ser	Lys	Val	Met	Ile	Ile	Glu	Asn	Ser	His	Val	Lys	Lys	1185	1190	1195
Asp	Asp	Ile	Trp	Pro	Ser	Gly	Gly	Gln	Met	Thr	Val	Lys	Asp	Leu	Thr	1205	1210	1215
Ala	Lys	Tyr	Thr	Glu	Gly	Gly	Asn	Ala	Ile	Leu	Glu	Asn	Ile	Ser	Phe	1220	1225	1230
Ser	Ile	Ser	Pro	Gly	Gln	Arg	Val	Gly	Leu	Leu	Gly	Arg	Thr	Gly	Ser	1235	1240	1245
Gly	Lys	Ser	Thr	Leu	Leu	Ser	Ala	Phe	Leu	Arg	Leu	Leu	Asn	Thr	Glu	1250	1255	1260
Gly	Glu	Ile	Gln	Ile	Asp	Gly	Val	Ser	Trp	Asp	Ser	Ile	Thr	Leu	Gln			

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 Gln Trp Arg Lys Ala Phe Gly Val Ile Pro Gln Lys Val Phe Ile Phe  
                                  1285                      1290                      1295  
 Ser Gly Thr Phe Arg Lys Asn Leu Asp Pro Tyr Glu Gln Trp Ser Asp  
                                  1300                      1305                      1310  
 Gln Glu Ile Trp Lys Val Ala Asp Glu Val Gly Leu Arg Ser Val Ile  
                                  1315                      1320                      1325  
 Glu Gln Phe Pro Gly Lys Leu Asp Phe Val Leu Val Asp Gly Gly Cys  
                                  1330                      1335                      1340  
 Val Leu Ser His Gly His Lys Gln Leu Met Cys Leu Ala Arg Ser Val  
 1345                      1350                      1355                      1360  
 Leu Ser Lys Ala Lys Ile Leu Leu Leu Asp Glu Pro Ser Ala His Leu  
                                  1365                      1370                      1375  
 Asp Pro Val Thr Tyr Gln Ile Ile Arg Arg Thr Leu Lys Gln Ala Phe  
                                  1380                      1385                      1390  
 Ala Asp Cys Thr Val Ile Leu Cys Glu His Arg Ile Glu Ala Met Leu  
                                  1395                      1400                      1405  
 Glu Cys Gln Gln Phe Leu Val Ile Glu Glu Asn Lys Val Arg Gln Tyr  
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 Asp Ser Ile Gln Lys Leu Leu Asn Glu Arg Ser Leu Phe Arg Gln Ala  
 1425                      1430                      1435                      1440  
 Ile Ser Pro Ser Asp Arg Val Lys Leu Phe Pro His Arg Asn Ser Ser  
                                  1445                      1450                      1455  
 Lys Cys Lys Ser Lys Pro Gln Ile Ala Ala Leu Lys Glu Glu Thr Glu  
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